**Glycine Intracerebroventricular Administration Impairs Energy Metabolism in Cerebral Cortex and Striatum of Young Rats**

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**Introduction:** Nonketotic hyperglycinemia (NKH) is an autosomal recessive inborn error of glycine (GLY) catabolism caused by a defect in GLY cleavage system and biochemically characterized by brain accumulation of this amino acid. Clinically, patients have spasticity, apnea, mental retardation and seizures that can lead to early death. Considering that the mechanisms of brain damage found in NKH are not fully established, the objective of the present study was to investigate the ex vivo effects of GLY administration on important parameters of energy metabolism in cerebral cortex and striatum of young rats. **Material and Methods:** Thirty-day-old Wistar rats were sacrificed 30 min after receiving a single intracerebroventricular injection of GLY (5 µmol) or NaCl (5 µmol). The cerebral cortex and striatum were dissected and homogenized in a specific buffer for each technique and used to measure CO₂ production from glucose, the activities of the respiratory chain complexes and creatine kinase (CK). **Results and Discussion:** Our results demonstrate that GLY significantly reduced CO₂ production from glucose in striatum, but not in cerebral cortex. GLY also decreased the activity of complex I-III and complex IV of the electron transfer chain in the striatum and in the cerebral cortex, respectively. We also observed that GLY inhibited the activity of total and mitochondrial CK in both brain structures. Furthermore, the compounds N-acetylcysteine and creatine prevented the effects of GLY on the activity of total CK in cerebral cortex and striatum, indicating the involvement of reactive species in these effects. **Conclusions:** The present data indicate that GLY impairs brain homeostasis at the level of energy transfer and formation. Therefore, it may be presumed that bioenergetic dysfunction contributes, at least in part, to the brain injury found in NKH.

**Keywords:** Nonketotic hyperglycinemia, glycine, energy metabolism, cerebral cortex, striatum.  
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